CPAM – Congenital Pulmonary Airway Malformation is a rare abnormal growth or mass of lung tissue that occurs when a fetus is developing in the womb. Until recently it was known as CCAM – Congenital Cystic Adenomatoid Malformation. The growth is usually confined to one lobe (section) of one lung. While it is not cancer, the tissue in the mass cannot function as normal lung tissue.

**Cause**
There is no known cause of CPAM. It is not related to genes (inherited) and there are no reported cases of it recurring in future pregnancies.

**Types**
CPAM is divided into types:

**Type I** This type of CPAM is usually made up of quite large cysts, but there tend to be few in number. The outcome for Type I is generally favorable.

**Type II** Type II often has a larger number of cysts but they are smaller in size. Type II is more likely to occur with other conditions or birth defects. These may include problems with the heart, kidneys, and skeletal or other body systems. The outlook for infants with Type II depends upon the severity of the other conditions.

**Type III** is the least common and most serious type of CPAM. They are usually large masses that can cause pressure on the heart, windpipe and lung on the other side causing them to shift to one side. When this occurs, the heart may not be able to function well. This can lead to hydrops – the accumulation of fluid in at least two of these areas: around the heart or lungs, under the skin, and/or in the abdomen.

**Diagnosis**

**During Pregnancy**
A mass in the lungs may be seen during an ultrasound exam. Further testing such as targeted ultrasounds will need to be done to confirm the diagnosis.

**After Birth**
If the CPAM was not detected during pregnancy, the baby may not show any signs at birth. If this is the case, CPAM may not be diagnosed until later in life or never be diagnosed.

Some babies with CPAM do have breathing problems and are diagnosed after birth. Breathing problems can affect heart rate and intensive care is needed.
CVR Monitoring
Once CPAM is detected, a CPAM volume ratio (CVR) is calculated using the size of the mass and the size of the baby's head. The CVR is a more precise way of measuring the CPAM growth.

CPAM masses may shrink in size, stay the same size, or grow in size. Frequent ultrasound tests (sometimes once or twice weekly) and CVR values are done to see if and how fast the mass is growing.

This CVR number will be followed closely to help determine treatment options. If the CVR value is above 1.6, there is a high risk of the baby developing heart and breathing problems and hydrops.

There is usually a point at which the mass stops or slows growing. This is called a plateau and occurs between 23 to 30 weeks of pregnancy – most often around week 25. A plateau may occur on its own or with treatment given to the mother.

Treatment
In many cases, the mass is small enough that the pregnancy can continue to birth without complications.

It is also possible – not likely – that a CPAM will shrink or disappear before birth. If this occurs, the baby will have imaging studies such a CT scan after it is born to confirm that it is no longer present.

The goal of treatment during pregnancy is to avoid or treat hydrops as soon it begins.

The treatment depends on the makeup (solid or cysts) of the CPAM, the CVR value, and whether there are signs of hydrops.

Steroid Medication
Steroid medication may be given to the pregnant woman to help slow or stop the growth. It is hard to know how effective this is since CPAM growth can slow on its own.

Cyst Aspiration/Shunt
If the CPAM has a large cyst, fluid may be removed with a needle. This is inserted through the mother's abdomen and the uterus to reach the mass in the fetus.

If the fluid recurs, a shunt (small hollow tube) may be inserted into the mass to continuously drain the fluid into the amniotic sac. Both of these procedures are done during pregnancy at hospitals specializing in this type of care.

Surgery
Surgery to remove the lobe of the lung with the mass may be performed during pregnancy if the CPAM is solid and there are signs of hydrops.

If the mass is large but there are no signs of hydrops, surgery may be done to the fetus followed by a C-section. This is called an EXIT (ex utero intrapartum treatment) procedure. It allows the surgery to be performed while the fetus is still attached to the umbilical cord resulting in better outcomes for the baby. This procedure and surgery during pregnancy are performed at specialized hospitals.
If the mass is not large, surgery can be performed after birth. Sometimes it is done within days. In other cases the baby may go home to grow and mature for a few weeks to a few months before having surgery.

Outlook

The long term outlook for infants after successful surgical removal of the CPAM is very good. Once the mass and lobe of the lung is removed, the remaining lung tissue will have extra growth to make up for the missing tissue. Children should be able to lead active normal lives.